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Giant intracranial arteriovenous malformation – a case report

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Summary

Intracranial arteriovenous malformations (AVMs) occur in 0.5-1% of the population [1]. An arteriovenous malformation is a tangled cluster of vessels, in which arteries connect directly to veins with no intervening capillary bed. Because an intracranial hemorrhage, or, rarely, a seizure are the first clinical symptoms of AVMs, they are the most dangerous congenital vascular malformations [1, 3, 4, 5].

We report a case of a 37-year-old woman with a giant intracranial arteriovenous malformation, who complained of headache attacks. The lesion was diagnosed in computed tomography and computed angiography of the head. The first imaging study in patients with suspected AVM is usually CT or angio-CT. These studies are good for depicting an AVM, and they are relatively noninvasive. However, angiography used for the diagnosis and planned treatment is invasive.

The authors present also the options for therapy.

Key words: Arteriovenous malformations • angio-CT option • spatial reconstruction

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Background

Intracranial arteriovenous malformations (AVMs), also referred to as arteriovenous hemangiomas, occur in ca. 0.5-1% of the population [1]. From the anatomopathological point of view, this abnormality involves direct connection of arteries with veins with no intervening capillary bed. Radiological assessment takes into consideration the location of AVMs, their size, shape, number, arterial blood supply, venous drainage, angioarchitecture and the presence of additional abnormalities, e.g. aneurysms. All these factors are important in the choice of appropriate treatment method. In 70-90% of cases, their location is supratentorial, in ca. 10% subtentorial [2]. Most often the anomalies of this type are localized in the temporal or parietal lobes. From the point of view of location, they are divided into superficial (gyric hemangiomas) deep (subgyric hemangiomas), midbrain and paraventricular ones. According to the source of blood supply, AVMs can be classified as:

- interstitial or leptomeningeal ones (75%) supplied by the internal carotid or vertebral arteries, which are in most cases congenital anomalies and become symptomatic in the second-fourth decade of life.
- dural ones (10%) supplied by the external carotid arteries, which are most frequently acquired malformations, often associated with a trauma or surgical procedure, which become symptomatic in the fourth-sixth decade of life.
- mixed ones (15%) with blood supply both from the branches of the external and internal carotid arteries [2].

The venous blood can be drained to the deep, or superficial venous system, or both. With respect to their size, the arteriovenous hemangiomas can be classified as:

- small (30%) with vascular cluster diameter < 3 cm,

- medium-sized (60%) with vascular cluster diameter of 3-6 cm,
- large (10%) with vascular cluster diameter > 6 cm. [2, 3]

Clinical manifestations of arteriovenous hemangiomas are most often observed in young adults, frequently in the 4th decade of life [3, 4, 5], with more or less equal incidence in males and females. In 41-79% of cases, they are manifested by an intracranial hemorrhage, the risk of which amounts to 2-3% / year. [1, 3, 5]. The risk of bleeding recurrence during the first year is 6% [1,3], and it should be remembered that each subsequent hemorrhage increases the risk of death by ca. 10-15% [2]. Other symptoms of intracranial arteriovenous hemangiomas include epileptic seizures – in 11-33% of cases (mainly in younger patients), headache (7-48%), progressive neurological deficits (1-40%), as well as ischemic disorders due to steal syndrome [3, 4, 5]. In view of the above, intracranial arteriovenous malformations belong to the most dangerous, life-threatening vascular anomalies, and therefore they should be diagnosed and treated as early as possible.

Case report

A 37-year-old woman was admitted to the gynecological department for treatment of large leiomyomata of the uterus. Anamnesis revealed that the patient had been for 7 years in charge of the neurology department because of an intracranial arteriovenous hemangioma located in the left parietooccipital region.. Since early childhood, the patient had complained of headaches associated with vomiting and right-sided numbness of the extremities. She had had the first episode of loss of consciousness and seizure about seven years before. Cranial CT performed at that time revealed an irregularly shaped anomaly containing single calcifications and showing intensive enhancement after contrast medium administration, which might correspond to an arteriovenous malformation, in the posterior part of the left cerebral hemisphere. Extension of diagnostics with

angiography was indicated. Angiography of cerebral arteries visualized a giant arteriovenous malformation, located in the left parietooccipital region, with blood supply from three sources:

- left posterior cerebral artery
- left anterior cerebral artery
- left middle cerebral artery.

Because of high surgical risk, neurosurgical treatment was abandoned and radiotherapy – 34.96 Gy in 19 fractions – was instituted. Because of the above findings, in order to assess the risk associated with the gynecological procedure, a consultation with a neurosurgeon was planned, as well as control angio-CT of the head. The imaging study was performed using a spiral one-row Somatom Emotion equipped set on brain angio-CT protocol, with 100 ml contrast medium administered i.v. with 3 ml/s flow velocity and 18 s delay. A massive lesion in the form of an arteriovenous hemangioma was visualized within the posterior cranial fossa in the parietooccipital region on the left.

The detected abnormality received blood supply from the Willis ring arteries, especially the left-sided ones (the middle and posterior cerebral arteries and branches of the basilar artery, and from the anterior cerebral artery via the commissural arteries). A distension of the cerebral venous system was also noted, particularly in the area of confluence of the sinuses and the sinus rectus.

Laboratory tests, such as blood cell count and biochemistry, as well as routine chest X-ray, revealed no significant abnormalities. After consultation with a neurosurgeon, considering the anamnesis data and radiological presentation, surgical treatment, both gynecological and neurosurgical, was abandoned. Only life-saving surgical procedures could be performed in the patient's condition.

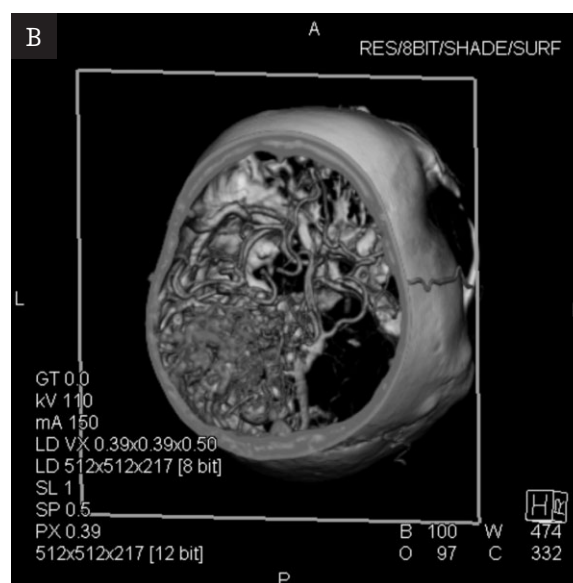
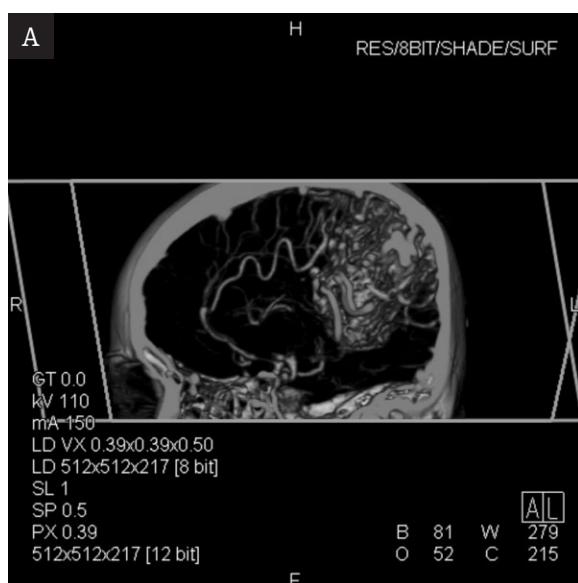


Figure 1 A, B. CT-angiography with spatial reconstruction. A giant intracranial arteriovenous malformation.

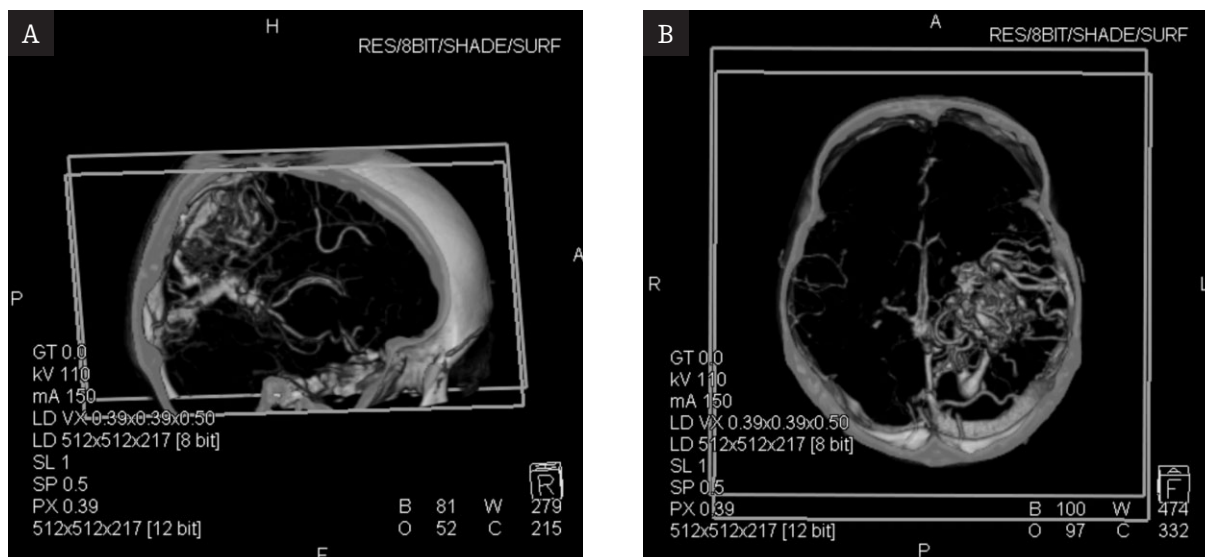


Figure 2 A, B. Arteriovenous malformation in angio-CT with spatial reconstruction, sagittal view (a), frontal view (B)

Discussion

The diagnosis of AVMs is based on the results of CT and MR, particularly with the angio option (angio-CT, MRA). Classic angiography, which was a „golden standard“ of diagnostics in pathologies of this type in quite recent times, is currently more and more often replaced by angio-CT with spatial reconstruction, performed with the use of multi-row CT scanners, the availability of which is increasing. Non-contrast-enhanced CT can visualize only indirect symptoms which may indicate the presence of AVMs: calcifications in the vascular walls or within the thrombi present in the hemangioma (detectable also in conventional skull X-ray), ischemic lesions and brain tissue atrophy in the nearest vicinity of the anomaly. After administration of contrast medium, numerous, distended, tortuous afferent and efferent blood vessels are visualized. Additional MRI examination is very helpful in assessment of the vascular damage area after radiosurgery, monitoring the condition of brain tissue surrounding the site (zones of edema or necrosis after treatment). However, the use of both CT and MR modalities is associated with certain limitations. Both in CT and in MRI, visualization of all small blood vessels can sometimes be impossible. It should also be mentioned that the presence of a hematoma formed as a result of injury, compressing the vascular cluster, makes it impossible to diagnose AVMs on the basis of the aforementioned investigations. Conventional angiography, which is a more invasive procedure, should be used in the cases in which embolization of the malformation during the procedure is planned. It allows to visualize the structure of the malformation in detail, with all the afferent vessels, as well as to assess its relation with the adjacent vascular structures. The data obtained as a result of radiological investigations provide the basis for classification of the hemangioma as belonging to one of surgical risk groups distinguished according to Spetzler-Martin scale. This score assesses 3 characteristic features of the lesion: size (0-3 cm: 1 point, 3-6 cm: 2, >6 cm: 3), deep venous drainage (absent – 0, present – 1) and location in relation to important centers of the brain (an area not associated with speech function – 0, an area not associated with speech function – 1). The

summary score determines the risk level: I° – 1 point; II° – 2 points, III° – 3; IV° – 4; V° – 5 [1, 3]. Three methods of treatment are currently applied in the cases of hemangiomas: surgical resection, percutaneous embolization and radiosurgery. To optimize treatment, they can be used in combinations, e.g. embolization before surgical resection [1]. The primary aim of percutaneous embolization is to reduce the malformation size, which can consequently shorten the duration of the surgical procedure and reduce intraoperative bleeding, or, in case of radiosurgery, to enable the use of a lower radiation dose [1]. Currently, surgical treatment is recommended for I-II° abnormalities according to Spetzler-Martin's classification. Also children and young adults are usually qualified for radical intervention. In patients from risk group III, combined treatment is used (embolization + surgery, embolization + radiosurgery). In patients belonging to groups IV and V, the risk of postoperative complications (irrespective of the treatment method) is so high that leaving the abnormality without any treatment with periodic control examinations should be considered [3, 6]. Radiosurgical techniques are applied in case of small lesions (<3 cm or larger if the procedure is combined with embolization), situated in the vicinity of important structures of the brain, where radical surgery would be associated with a high risk of complications [1, 6]. Each patient diagnosed with an arteriovenous malformation should be assessed individually to determine the benefits of surgical treatment and the risk of potential complications. In the reported case, the patient was not qualified for radical treatment. Surgical intervention in case of hemangiomas located in the parietal region supplied by the branches of the anterior cerebral artery, the commissural and frontal artery, is often connected with loss of motor function of the limbs [1]. Sometimes the best solution is to leave the abnormality without treatment with periodic control examinations. The annual hemorrhage risk rate is 2-4% in such cases [1].

Conclusions

1. Angio-CT is a very good, low-invasive diagnostic method and should be used as the first line examination in case of suspected arteriovenous malformation.

2. Angiography, which is more invasive and more burdensome for the patient, should be used if therapeutic surgery is planned.
3. Each patient diagnosed with an arteriovenous malformation should be assessed individually to determine the benefits of surgical treatment and the risk of potential complications.

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